

What is claimed is:

1. A polynucleotide comprising a mutant of the nucleotide sequence coding for the factor VII-activating protease (FSAP), comprising at least one of a G to C base exchange at nucleotide position 1177 and a G to A base exchange at nucleotide position 1601.
2. The polynucleotide as claimed in claim 1, comprising the nucleotide sequence of SEQ ID NO:2.
3. A polypeptide comprising an FSAP mutant, having at least one of a Glu to Gln exchange at amino acid position 393 and a Gly to Glu exchange at amino acid position 534.
4. The polypeptide as claimed in claim 3, comprising the amino acid sequence of SEQ ID NO:4.
5. A diagnostic method for identifying genetic heterozygous or homozygous expression of the FSAP mutant according to one of claims 3 or 4, wherein a polynucleotide encoding the FSAP mutant, or encoding a proenzyme or fragment thereof is detected.
6. A diagnostic method for identifying genetic heterozygous or homozygous expression of the FSAP mutant according to one of claims 3 or 4, wherein a polypeptide comprising the FSAP mutant or a proenzyme or fragment thereof is detected.
7. A monoclonal or polyclonal antibody, which is specifically directed against at least one of FSAP, and proenzymes and fragments thereof, or specifically directed against at least one of FSAP mutants containing at least one of a Glu to Gln exchange at amino acid position 393 and a Gly to Glu exchange at amino acid position 534, and proenzymes and fragments thereof.

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8. A diagnostic method, which comprises detecting at least one of FSAP, its proenzyme, its fragments, and its mutants by using a monoclonal or polyclonal antibody as claimed in claim 7.
9. The diagnostic method as claimed in claim 8, which comprises one of
 - a) incubating a sample which could contain an FSAP mutant polypeptide with a first antibody fixed to a solid support, then, after washing, adding a second, labeled antibody and, after washing out again, measuring the signal produced by the second antibody, wherein the first and second antibodies are specific for at least one of FSAP, and proenzymes and fragments thereof, or specific for at least one of FSAP mutants containing at least one of a Glu to Gln exchange at amino acid position 393 and a Gly to Glu exchange at amino acid position 534, and proenzymes and fragments thereof;
 - b) incubating a sample which could contain an FSAP mutant polypeptide with a first antibody fixed to a solid support and directed against the FSAP wild type, then, after washing, adding a second, labeled antibody specific for at least one of FSAP, and proenzymes and fragments thereof, or specific for at least one of FSAP mutants containing at least one of a Glu to Gln exchange at amino acid position 393 and a Gly to Glu exchange at amino acid position 534, and proenzymes and fragments thereof, and, after washing out again, measuring the signal produced by the second antibody;
 - c) fixing the sample to be tested for the presence of an FSAP mutant polypeptide to a support and detecting said sample with a labeled antibody specific for at least one of FSAP, and proenzymes and fragments thereof, or specific for at least one of FSAP mutants containing at least one of a Glu to Gln exchange at amino acid position 393 and a Gly to Glu exchange at amino acid position 534, and proenzymes and fragments thereof alone or in a mixture with an unlabelled antibody and subsequent detection of the labeled antibody; and
 - d) adding a sample to be tested for the presence of an FSAP mutant polypeptide to an antibody specific for at least one of FSAP, and proenzymes and fragments thereof, or specific for at least one of FSAP mutants containing at least one of a Glu

to Gln exchange at amino acid position 393 and a Gly to Glu exchange at amino acid position 534, and proenzymes and fragments thereof fixed to a support, in the presence of a labeled FSAP mutant and measuring the signal produced by the label.

10. The diagnostic method as claimed in claim 8, wherein FSAP activity is measured by
 - a) incubating the sample containing one or more of FSAP wild type, FSAP mutants, FSAP proenzymes, and FSAP fragments on a solid support to which an antibody specific for at least one of FSAP, and proenzymes and fragments thereof, or specific for at least one of FSAP mutants containing at least one of a Glu to Gln exchange at amino acid position 393 and a Gly to Glu exchange at amino acid position 534, and proenzymes and fragments thereof has been coupled beforehand; and
 - b) after washing out the free support, incubating the protein fixed to said support with reagents which allow determination of its activity.
11. The method as claimed in claim 8, wherein the antibody is used for detecting said at least one of FSAP, its proenzyme, its fragments, and its mutants by Western Blots, immunohistology, or fluorescence-assisted cell sorting (FACS).
12. A method for preparing the polypeptide as claimed in one of claims 3 or 4, which comprises fixing one or more antibodies specific for at least one of FSAP, and proenzymes and fragments thereof, or specific for at least one of FSAP mutants containing at least one of a Glu to Gln exchange at amino acid position 393 and a Gly to Glu exchange at amino acid position 534, and proenzymes and fragments thereof to a support, incubating the immunoadsorbent with the sample containing the polypeptide and then, after washing, obtaining the polypeptide by elution.
13. The method as claimed in claim 12, wherein polypeptides are expressed by at least one of recombinant and transgenic expression.

14. The method as claimed in claim 12, wherein the polypeptide is obtained from a source selected from body fluids, cell culture supernatants, and fluids of transgenic animals.
15. A method of administering the polypeptide of one of claims 3 or 4, or an antibody directed against said polypeptide, or a combination of said polypeptide and said antibody for prophylactic or therapeutic inhibition of bleeding in the case of congenital or acquired deficiency in one or more of FVIII von Willebrand factor, FV, FIX, FX, FXI, and FXII.
16. A monoclonal antibody against FSAP, FSAP mutants, FSAP proenzymes or FSAP fragments, which is produced by the hybridoma cell line DSM ACC2453 or hybridoma cell line DSM ACC2454.
17. A method for purification, detection, and optionally activity determination of FSAP, its mutants, its fragments, or its proenzyme, wherein the monoclonal antibody as claimed in claim 16, its Fab or its F(ab')₂ fragment is used.
18. A diagnostic method for detecting antibodies against at least one of FSAP and an FSAP mutant formed by the exchange of one or more amino acids, which comprises letting a sample which could contain the antibodies react with at least one of FSAP and FSAP mutants which are fixed to a solid support and, after washing, detecting the antibodies bound to the support.
19. The diagnostic method as claimed in claim 18, wherein the antibodies bound to the solid support are incubated with a substance selected from labeled anti-human immunoglobulin or fragments thereof, labeled protein A, and protein G, and wherein the signal emitted by the labeled substance bound to the support is determined.
20. The diagnostic method as claimed in claim 18, wherein the antibodies are detected by photometric measurement of the extinction caused by cleavage of a suitable chromogenic or fluorogenic substrate by anti-human immunoglobulin or fragments thereof, protein A, or protein G bound to the support.

21. The diagnostic method as claimed in claim 18, wherein the antibodies are detected by fluorescence measurement.
22. The diagnostic method as claimed in claim 18, wherein the antibodies are detected by radiometric measurement.
23. The diagnostic method as claimed in claim 8 for the immunohistological detection of at least one of FSAP, its proenzyme, its fragments and its mutants, which comprises letting an anti-protease, labeled, monoclonal or polyclonal antibody or one of its fragments react with a tissue sample, and washing out the unbound antibody or its fragments and determining the signal emitted from the bound antibody or one of its fragments.
24. The diagnostic method as claimed in claim 8, for the immunohistological detection of at least one of FSAP, its proenzyme, its fragments and its mutants, which comprises letting an unlabelled monoclonal or polyclonal antibody, directed against FSAP, or a proenzyme, mutant, or fragment thereof react with the tissue sample, washing out the unbound antibody or its fragments, then letting a labeled anti-antibody react with the tissue and, after washing out the unbound labeled anti-antibody, determining the signal emitted from the bound anti-antibody or its fragments.
25. The diagnostic method as claimed in one of claims 23 or 24, wherein the tissue sample to be tested is taken from an endocrine organ.
26. The diagnostic method as claimed in one of claims 23 or 24, wherein the antibodies comprise monoclonal antibodies of hybridoma cell lines DSM ACC2453 or DSM ACC2454.
27. An assay system with which the method as claimed in claim 5 is carried out.
28. An assay system with which the method as claimed in claim 6 is carried out.

29. An assay system with which the method as claimed in one of claims 8, 9, 10, 11, 23, or 24 is carried out.

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